Supratentorial intra-arachnoid cyst

Case report

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Unlike the cases of intra-arachnoid cyst reported previously, this case illustrates the clinical and pathological evidence of progressive expansion of the cyst, eventually resulting in a fatal outcome, and provides further pathological evidences to indicate that intra-arachnoid cysts may constitute a distinct pathological entity among the heterogeneous cysts overlying the cerebral hemispheres.1−6

Case Report

In February, 1969, a 72-year-old woman was admitted to the hospital after 2 hours of right-sided seizures. In 1962 she had experienced the insidious onset of mild right-sided weakness and dysphasia, which was initially attributed to a “stroke” but which was slowly progressive over the next 6 years. About 6 months before admission, she began having right-sided seizures, rapid progression of right hemiparesis and dysphasia, and occasions of falling. On the day of admission, she fell striking her head without loss of consciousness and 10 minutes later had the right-sided seizure which precipitated her admission to Morrisania City Hospital.

Examination. The patient had no external signs of head trauma and 12 hours after admission was alert and able to follow simple commands. She was expressively aphasic and had a right hemiparesis including the face and a depressed right corneal reflex. Lumbar puncture revealed normal fluid with an opening pressure of 180 mm H2O. The electroencephalogram (EEG) was diffusely slow with a left frontotemporal delta focus. The brain scan was negative.

She did well for 1 day but then became increasingly lethargic and was transferred to Montefiore Hospital and Medical Center for further evaluation. En route she developed bilaterally dilated and fixed pupils and required tracheal intubation to assist respiration. Upon arrival, she responded to deep pain with triple flexion of the right leg; she had fixed dilated pupils, normal ocular fundi, a flattened right side of the face, and...
FIG. 1. Superior view of the left cerebral hemisphere showing the large cyst roofed by a semitranslucent membrane (A) continuous with the arachnoid membrane covering the hemisphere and distinct from the overlying dura (D). The indented brain shows flattened gyri and shallow sulci containing blood vessels.

absent oculocephalic and cold caloric responses. There was left-sided flaccidity and increased tone on the right. There was bilateral ankle clonus. An emergency left carotid angiogram demonstrated a large avascular mass extending superficially and also involving a considerable thickness of the left cerebral hemisphere in the Rolandic area. There was a gyral blush posterior to the avascular zone. There was no bulging or thinning of the calvarium. She was treated with parenteral steroids and oral glycerol but died 2 days after admission.

Autopsy Findings

The autopsy was performed at the medical examiner’s office of the City of New York and the brain was obtained from there preserved in formalin. The brain weighed 1300 gm. On external examination, a large cyst was seen on the convex surface of the left cerebral hemisphere occupying the posterior part of the frontal and anterior third of the parietal lobes (Fig. 1). The cyst measured $8 \times 4 \times 4$ cm and extended medially to within 2 cm of the midline. The central fissure as well as the adjacent sulci were incorporated in the cyst and could not be identified. The roof of the cyst was formed by a thin semitranslucent membrane that was distinct from the overlying dura and appeared to be continuous with the arachnoid membrane covering the surface of the adjacent brain. The roof was partially torn during removal of the brain. The cyst had contained clear fluid which was not available for chemical study. No evidence of hemorrhage or adhesion was seen in the vicinity of the cyst. The cyst in its deeper part was limited by the markedly depressed surface of the brain, which had flattened gyri.

On coronal sections, the deeper extent of the cyst as well as the severe deformity of the brain resulting from it were more clearly defined (Fig. 2). Despite the marked reduction in width, the cortical ribbon of the indented brain was intact. The subjacent white matter of the centrum semiovale was compressed to some extent. The Sylvian fissure was compressed and displaced downward. The cyst at no point communicated with the ventricle. At its closest approximation to the ventricle, the deepest part of the cyst was separated from the angle of the lateral ventricle by at least 1 cm of white matter. In addition to the local indentation, the brain was markedly distorted by the cyst with resultant herniations of the left cingulate gyrus and the hippocampus. There were a massive secondary brain stem hemorrhage and fresh infarcts involving both frontal lobes, the left occipital lobe, and the basal ganglia (Fig. 2). There was a moderate degree of cerebellar tonsillar herniation.

Multiple specimens representing the peripheral and deeper parts of the cyst wall, including those from the edge of the cyst, were embedded and sections were stained with hematoxylin and eosin, Nissl, Woelcke, Holzer, and Wilder’s reticulin stains. Additional samples from the rest of the brain were prepared in a similar manner. Microscopically, the membranous roof of the cyst had an appearance identical to that of the arachnoid membrane and consisted of a thin layer of connective tissue lined by flattened cells. The cyst wall that had been adjacent to the indented brain was enclosed by a similar membrane which, after fusing with the membran-
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nous roof at the edge of the cyst, was continuous with the arachnoid membrane covering the surface of the hemisphere. The enclosure of the cyst by the duplicated arachnoid membrane was further evidenced by the fact that the inner arachnoid membrane was distinctly separated from the pia mater by a subarachnoid space containing the usual number of leptomeningeal blood vessels (Fig. 3).

Despite marked narrowing, the subarachnoid space was discernible in most sections. In the deepest part of the cyst, however, the subarachnoid space was virtually obliterated except for the space occupied by the leptomeningeal blood vessels (Fig. 4). Occasionally, these protruded into the cyst, ensheathed by folds of the inner arachnoid membrane in a manner analogous to the mesentery of the peritoneum (Fig. 4 inset). Elsewhere in the deeper part of the cyst, the inner membrane was closely apposed to the pia mater. Due to partial collapse of the cyst, the inner arachnoid membrane became redundant to a variable extent, particularly toward the edge of the cyst (Fig. 5).

The lining cells of the inner arachnoid membrane were, in general, plumper than those lining the normal arachnoid membrane (Fig. 5). However, these cells were surrounded by delicate reticulin fibers, similar to the lining cells of the leptomeninges elsewhere (Fig. 3). There was no evidence of old hemorrhage, fibrosis, or infection anywhere in the cyst wall. The indented cerebral

Fig. 2. Coronal sections of the brain showing the deeper extents of the cyst and severe herniations of the left hippocampus (arrows) and the cingulate gyrus. The cortical ribbon of the indented brain is markedly reduced in width. Note hemorrhage in the midbrain and fresh infarction in the region of the parietal lobe and basal ganglia.

Fig. 3. Microscopic section of the cyst wall adjacent to the indented brain showing a thin arachnoid membrane (A) predominantly composed of connective tissue which separates the cystic cavity (C) from the compressed cerebral cortex with normal pial covering (P). Note the subarachnoid space containing the usual number of blood vessels. Wilder reticulin stain, × 70.

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FIG. 4. Microscopic section of the deeper part of the indented brain showing virtual obliteration of the subarachnoid space, except for the locations occupied by the blood vessels (SA) with occasional ensheathment by the inner arachnoid membrane (inset). Note severe atrophy and disorganized laminar pattern of the indented cerebral cortex. Celloidin H & E, × 34 (inset, × 136).

cortex showed some loss of neurons and a mild degree of reactive gliosis. The remaining neurons were arranged in a disorganized manner with resultant loss of normal laminar pattern (Fig. 4).

The terminal changes in the brain were even more extensive and consisted of widespread fresh infarcts and secondary brain stem hemorrhage. The corticospinal and other long tracts at various levels of the brain stem were of normal size and symmetrical with no detectable degenerative changes. There was a mild degree of segmental arteriosclerosis of the cerebral vessels.

Discussion
The formation of leptomeningeal cysts following trauma, hemorrhage, or infection is generally considered to be the result of localized entrapment of cerebrospinal fluid due to adhesions between the pia-arachnoid membrane. The pathogenesis of primary arachnoid cysts, however, is poorly understood. This may be, to a large extent, due to the lack of adequate histological study of various parts of the cyst wall, since the description of the clinically significant cysts of this variety is frequently based on observations at surgical exploration. The histological description of the cyst wall, when available,
Supratentorial intra-arachnoid cyst has been limited to its outer membranous portion. Due to the resemblance of this membrane to the arachnoid and apparent lack of an underlying cause, such cysts have been designated as primary arachnoid cysts. While mild trauma, usually unrecognized, is considered by some as an etiological factor in some instances, primary arachnoid cysts are generally assumed to be congenital in origin. Furthermore, Robinson in reporting a series of supratentorial cystic lesions with detailed review of the previously reported similar cases, including those located within the arachnoid membrane as reported by Starkman, et al., concluded that, in most instances, the so-called primary arachnoid cysts overlying the cerebral hemisphere, in reality, represent localized enlargement of the subarachnoid space due to agenesis of the underlying brain. Thus, such cystic spaces are in communication with the subarachnoid space and rarely cause compression or distortion of the brain unless complicated by loculation as a result of hemorrhage or adhesions.

The cyst in our case, however, was located within the arachnoid membrane rather than in the subarachnoid space. This was indicated by the fact that both outer and inner membranes enveloping the cyst fused with each other at the edge and continued as the normal arachnoid membrane covering the surface of the adjacent brain. Simultaneously, the inner arachnoid membrane maintained a normal relationship to the pia mater overlying the severely indented brain. Furthermore, continued expansion of the cyst in this patient was clearly evidenced, not only by the progressive neurological deficits terminating in a fatal outcome, but by the severe pathological alterations of the brain due to mechanical compression and raised intracranial pressure, especially in the absence of such complicating factors as hemorrhage or adhesions. This cyst, therefore, probably does not represent an enlargement of the subarachnoid space due to agenesis of the underlying brain. Moreover, the normal development of the corticospinal tract on the affected side emphasizes the unlikelihood of agenesis.

The intra-arachnoid location of the cyst as described here was basically similar to the six cases studied in detail at autopsy. Interestingly, all six patients were adults. Unlike the present case, however, the cysts in those cases were located in the region of the Sylvian fissure and in all but one instance were asymptomatic. Moreover, marked alterations of the brain resulting from progressive expansion of the cyst such as those described in the present case were lacking in those cases.

The demonstration of the precise location of the cysts within the arachnoid membrane by means of detailed histological study at autopsy appears to be important particularly in view of the obscure nature of primary arachnoid cysts. While in some instances such cysts may indeed be the result of localized enlargement or loculation of the subarachnoid space, it appears that the intra-arachnoid cysts constitute a group of supratentorial arachnoid cysts pathologically distinct from those ascribed to trauma or cerebral agenesis. The focal duplication of the otherwise thin arachnoid membrane enclosing such cysts would further indicate the possi-
bility of an underlying developmental defect. Starkman, et al., suggested that the intra-arachnoid cysts may be the result of a focal derangement of the normal mechanism by which the leptomeninges are derived from the perimedullary mesenchyma. A false passage in the developing leptomeninges created by the pumping force of the cerebrospinal fluid could conceivably be closed off from the subarachnoid space, thus forming a cyst. The mechanism of the progressive enlargement of this kind of a cyst still remains unexplained.

The true incidence of intra-arachnoid cysts is not known, although primary arachnoid cysts, in general, are frequently assumed to occur within the arachnoid membrane on the basis of limited histological examination of the cyst wall. The clinical features of the primary arachnoid cysts overlying the cerebral hemisphere appear to be quite variable. In some instances, these cysts remain silent throughout life and are discovered at autopsy as incidental findings. When symptomatic, such cysts may present themselves as space-occupying lesions with a variety of neurologic deficits. Frequently, the only manifestation of these cysts is limited to a local bulging of the overlying skull. Such a cyst usually appears on x-ray films as an avascular space-occupying mass. In children, in particular, additional changes are seen in the overlying skull characterized by local bulging and thinning.

Although rare in the adult, the possibility of a primary arachnoid cyst should be included in the differential diagnosis of slowly progressing supratentorial mass lesions. Early recognition and appropriate surgical treatment are important despite the usual benign course, since this potentially remediable lesion may otherwise result in irreparable brain damage or even fatal outcome, as indicated by the case reported here.

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References


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